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ATRIAL PACING: AN ALTERNATIVE TREATMENT FOR PROTEIN-LOSING ENTEROPATHY AFTER THE FONTAN OPERATION

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Protein-losing enteropathy (PLE) is an uncommon but potentially debilitating and difficult to manage complication after the Fontan operation. A number of treatment strategies, including diuretics, afterload reduction, intravenous albumin replacement, corticosteroids, heparin, creation of a fenestration, or revision of the Fontan pathway, have all been attempted, with various degrees of success.[1-4] Baffle fenestration is believed to augment cardiac output and increase oxygen delivery while lowering central venous pressure. Resolution of PLE after fenestration creation suggests that improvement in cardiac output and oxygen delivery are the key elements necessary for successful treatment of this disorder.

Another complication noted after the Fontan operation is sinus node dysfunction.[5] The presence of junctional rhythm and loss of atrioventricular synchrony can have deleterious effects on Fontan physiology by diminishing cardiac output. We report our experience with atrial pacing as treatment for PLE in patients undergoing the Fontan operation who manifest sinus node dysfunction.

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Clinical summaries

PATIENT 1. The patient underwent a nonfenestrated lateral tunnel Fontan operation for tricuspid atresia, ventricular septal defect, and pulmonic stenosis without complications at 2 years of age. Four years later, the patient had PLE with ascites and diarrhea (total protein, 4.3 g/dL; albumin, 2.5 g/dL; 24° stool α-1 antitrypsin clearance, 166 mL).* Temporary improvement occurred with diuretics, supplemental albumin infusions, and corticosteroids. The patient was unable to be weaned off of steroids after a return of PLE. Cardiac catheterization revealed a mean right atrial pressure of 13 mm Hg, an unobstructed systemic venous pathway, and good ventricular function. One year later, the patient underwent surgical creation of a single 4.8-mm fenestration. The oxygen saturation decreased to 84% with resolution of PLE (total protein, 7.2 g/dL; albumin, 4.1 g/dL). An ambulatory monitor 2 years later revealed a predominant junctional rhythm at 65 beats/min. The patient continued to do well (oxygen saturation, 85%) on

*Normal laboratory values: total protein, 5.9 to 7.0 g/dL; albumin, 3.5 to 5.2 g/dL; and 24° stool α-1 antitrypsin clearance, 27 mL or less.

minimal diuretics and afterload reduction until the following year, when there was return of diarrhea, ascites, and periorbital edema (total protein, 3.7 g/dL; albumin, 1.8 g/dL). Repeat cardiac catheterization identified mild narrowing in the Fontan baffle, which was corrected with stent placement. With temporary atrial pacing in the cardiac catheterization laboratory, the systemic blood flow increased from 2.8 to 3.2 $L \cdot min^{-1} \cdot m^{-2}$. Symptoms of PLE persisted despite correction of the anatomic baffle narrowing. The patient subsequently underwent placement of an epicardial single-chamber pacemaker (Medtronic Thera 7960, Medtronic, Inc, Minneapolis, Minn) programmed at AAI 80 pulsations/min. Within 3 weeks, there was complete resolution of PLE. Six months later, the patient continues to be well without any edema or diarrhea (total protein, 7.2 g/dL; albumin, 4.3 g/dL).

PATIENT 2. The patient was admitted with hypoproteinemia, edema, and ascites 11 months after an extracardiac fenestrated Fontan operation for hypoplastic left heart syndrome (total protein, 4.4 g/dL; albumin, 2.1 g/dL; and 24° stool α-1 antitrypsin clearance, 52 mL). Temporary relief was achieved with intravenous diuretic therapy and albumin replacement. The patient was readmitted twice within a 12-month span for 25% albumin infusions. Intra-atrial re-entrant tachycardia was observed within the first few months after the Fontan operation. Adequate control of the arrhythmia was achieved on a combination of digoxin and amiodarone; however, marked junctional bradycardia followed 6 months later. Cardiac catheterization revealed a right atrial pressure of 10 mm Hg, an end-diastolic pressure of 5 mm Hg, and an arterial oxygen saturation of 96%. The cardiac index was 3.2 $L \cdot min^{-1} \cdot m^{-2}$, with no evidence of conduit obstruction or pulmonary artery distortion. Because of persistent sinus node dysfunction, a single-chamber epicardial pacemaker was implanted (AAI at 80 pulsations/min; Medtronic Kappa 701) 2 years after the Fontan operation. Within 3 weeks, the diarrhea resolved, and the patient has remained free of ascites, diarrhea, and peripheral edema (total protein, 7.2 g/dL; albumin, 4.4 g/dL).

Discussion. Enteric loss of proteins with the clinical findings of diarrhea, edema, ascites, immunodeficiency, and/or hypercoagulabilty is diagnosed with increasing frequency after the Fontan operation. The exact pathophysiologic mechanism is unknown, but it is likely related to a hemodynamic derangement of the intestinal circulation secondary to the Fontan physiology. As is typical in other patients with PLE, the 2 patients in this report presented at variable times after the Fontan operation (range, 1-4 years). Although both patients had some initial improvement with diuretic therapy and afterload reduction, symptoms of PLE returned. At our institution, all patients with PLE undergo a cardiac catheterization to exclude baffle obstruction or significant pulmonary artery distortion as a cause for elevated systemic venous pressure. One patient in our series had a narrowing in the Fontan baffle that was addressed with a stent; however, the clinical features of PLE persisted until an atrial pacemaker was implanted.

The resolution of clinical and laboratory PLE in these 2 patients after atrial pacing is likely a result of augmented cardiac output. Sinus node dysfunction may limit cardiac output through loss of the atrial contribution to ventricular filling. Although the results of a surgically created fenestration in the Fontan baffle have been acceptable with regard to PLE, it may leave the patient prohibitively cyanosed. Atrial pacing is an alternative way to increase cardiac output without the hypoxemia associated with the fenestration. The 2 patients in our report had sinus node dysfunction in addition to PLE. It is unknown whether other patients with PLE, but without sinus node dysfunction, would similarly benefit from the salutary effects of atrial pacing and increased cardiac output. Atrial pacing should be considered as part of the treatment regimen for patients with PLE after the Fontan operation.

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